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Pulmonary Hypertension

INCIDENCE AND MORTALITY OF PULMONARY HYPERTENSION ARE SIGNIFICANTLY RAISED BY COMBINATION OF PULMONARY INTERSTITIAL FIBROSIS WITH EMPHYSEMA

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Background: Combined pulmonary fibrosis and emphysema (CPFE) is a recently proposed syndrome consisting of upper lobe emphysema and lower lobe fibrosis in the lung. The aim of this study was to characterize impacts of CPFE on incidence and mortality of pulmonary hypertension (PH) in comparison with those of isolated intestinal lung disease (ILD).

Methods: Consecutive 108 subjects with interstitial lung disease who underwent both high resolution CT scan (HRCT) of the chest and transthoracic echocardiography were enrolled. After exclusion of 4 patients for poor images of pulmonary fibrosis, we divided 104 subjects into a group with CPFE group (n=41) and a group without CPFE (non-CPFE group, n=63). The clinical characteristics, pulmonary functions, ECG, echocardiographic data and prognosis were analyzed and compared between the two groups.

Results: In the CPFE group, 68.3% of the patients were male and 76.3% were current smoker or ex-smoker. The CPFE group had significantly higher estimated right ventricular systolic pressure (41.6 mmHg vs. 33.1 mmHg,P<0.01), lower FEV1% predicted (P=0.04), FVC% predicted (P=0.06), and percent diffusion lung capacity for carbon monoxide (DLCO%) (P=0.04) as compared with those in the non-CPFE group. Logistic regression analysis indicated that male gender and DLCO% were independent factors associated with CPFE. Kaplan-Meier survival curves showed that the CPFE patients had significantly higher mortality than the non-CPFE patients (P=0.015). In the CPFE group, logistic analysis indicated that voltage of R wave at V1 lead (V1R) is a predictor of death, and receiver operating characteristic analysis indicated that V1R > 0.5 mV predicts mortality with sensitivity of 80% and specificity of 70% (area under curve: 0.72). Difference in mortality between the subgroup with V1R > 0.5 mV and the subgroup with V1R < 0.5 mV in the CPFE group was confirmed by Kaplan-Meier survival curve analysis (P=0.031).

Conclusions: As compared with isolated ILD, CPFE is more frequently associated with PH and worse prognosis. The mortality of CPFE is predicted by high V1R, indicating contribution of PH to the poor prognosis of this syndrome.